

### Case report

## **Hereditary woolly hair- a rare case report**

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**Abstract:** Woolly hair is a rare congenital structural abnormality of the scalp hair manifesting as short, tightly coiled hair which may also involve the hairs over other parts of the body. We herein presents a case of 13 year old girl with scanty, extremely curly, coarse textured hairs over scalp like negroid.hairs since birth. Hairs on other body parts were normal. No family history was present. The case was reported due to its rarity of occurrence.

Keywords: Woolly hair, Keratosis pilaris

### **Introduction**

Woolly hair , also known as unruly hair, is extremely tangled and curly hair in an individual of non – negroid origin covering the whole scalp or in part. The scalp skin is clearly discernible in this.<sup>1</sup>

### **Case report**

A 13 year old girl presented with abnormal hair over scalp since birth( figure 1 and figure 2). There was no parental consanguinity. She was born out of full term normal delivery. Her developmental milestones were normal. No family history of similar disorder was present. Parents gave history that the scalp hair were sparse and curly since birth. The child had undergone shaving of the whole scalp twice at the age of 2 and 5 years however the hairs continued to grow curly and short.

On examination, the scalp hair were scanty, extremely curly, coarse textured and evenly pigmented like a negroid hairs (figure 1 and figure 2) . However, the hairs were not fragile or easily pluckable. Hairs on other body parts were normal. Rest of the cutaneous and systemic examination was unremarkable.

All the routine laboratory investigations were within the normal limits. Examination of the hair under light microscope did not reveal any abnormality except for the curling of the hair. Based upon all these details a diagnosis of hereditary woolly hair was made.

### **Discussion**

The term woolly hair has been derived from sheep wool.<sup>2</sup> Hutchinson et al<sup>3</sup> has classified woolly hair syndromes into four types. They are - Hereditary woolly hair, familial woolly hair, woolly hair



Figure 1:



Figure 2:

naevus and acquired progressive kinking of the hair. Hereditary woolly hair is inherited as autosomal dominant trait whereas familial woolly hair is inherited as autosomal recessive trait. In both of these a diffuse or generalized scalp involvement is seen. In woolly hair naevus, which is a localized form, woolly hairs appear well demarcated from the surrounding field of normal hairs . All the three disorders usually manifest at birth. On the other hand, APKH is a progressive change in the texture of certain scalp hair to a coarse, wiry or kinky state, occurring at or after

puberty. It is mainly localized to frontal and vertex regions of the scalp.<sup>2</sup>

Two characteristic associations of woolly hair are Naxos disease and Carvajal disease. Naxos disease is characterised by woolly hair, palmoplantar keratoderma and dilated cardiomyopathy with right ventricular dysplasia.<sup>4</sup> It is an autosomal-recessive disorder occurring due to mutation in the plakoglobin gene.<sup>5</sup> Carvajal disease is similar clinically to Naxos disease, except for left ventricular involvement and presentation at a younger age, and it is due to mutation in the desmoplakin gene.<sup>6</sup>

Woolly hair has been reported in association with keratosis pilaris atrophicans and cataract<sup>7</sup>, keratosis pilaris, curling of eyelashes<sup>8</sup>. A dominant form of woolly hair has also been reported in an English family with associated ichthyosis and deafness.<sup>9</sup> Additional features in a Dutch family with woolly hair included dental caries, acral keratoderma and facial abnormalities.<sup>10</sup> A case of woolly hair with keratosis follicularis spinulosa decalvans, teeth abnormalities, scarring alopecia and ophthalmological abnormalities has also been reported. The dental abnormalities included agenesis, inclusions and malformed teeth.<sup>11</sup> Another case of woolly hair associated with pachyonychia congenita, nail changes in the form of yellowish brown hypertrophy and wedge-shaped subungual hyperkeratosis of all 20 nails with natal teeth has been reported.<sup>12</sup> We reported the case due to its rarity of occurrence.

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